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ACUTE ANTERIOR POLIOMYELITIS IN A YOUTH.*

BY THEODORE A. HOCH, M.D.,
ASSISTANT PHYSICIAN AND PATHOLOGIST, WORCESTER INSANE HOSPITAL,
WORCESTER, MASS.

The case to be described is one of sporadic acute anterior poliomyelitis in a boy, 16 years of age, who died 13 weeks after the onset of the disease. Dr. B. T. Burley, of Worcester, observed the case and kindly furnished me with the clinical data.

On August 22nd, 1904, Dr. F. H. Baker called Dr. Burley in consultation to see the following case. H. S., a boy in his 16th year, was a member of a generally healthy family. One sister, however, two years older, had died three months before of tubercular peritonitis; and an older brother had had an ankylosed (tubercular?) knee since childhood. H. S. had always been especially healthy and had grown larger than any of the other children. Five days ago he did not feel as well as usual though he had no pain; felt languid and asked to be excused from doing the chores. The next day he had some headache and alternating hot and cold sensations. The headaches became more severe in the next two days and were accompanied by marked stiffness of the neck. The following night he arose to go to the bathroom and on getting on his feet slumped to the floor. It was found he could not walk and he was carried to his bed. On attempting to pass his urine at this time he failed and he was catheterized by the attending physician. The next morning it was found that the left shoulder could not be raised and the hand used but little. Later in the day the right arm also became weak. At the time of the physician's visit the next day he was told the temperature had been varying from 99.5 to 102.5 degrees, and that there had been at times slight delirium in the last two nights. Three physicians had already seen the boy and the prevailing diagnosis was cerebro-spinal meningitis.

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* Read at a meeting of the New England Psychological Society at Foxborough, Mass., March 28, 1905.

Physical Examination.—A well-developed and well-nourished boy. Face flushed. Skin rather hot and dry. Tongue coated and dry. Pupils equal and react normally to light and distance. Head normal, except for slight retraction and increased tension of the sub-occipital muscles. Heart not enlarged; no murmurs. Pulse 94. Abdomen not abnormal.

Muscular power: Patient could raise neither shoulder. Could use forearm and biceps on right; flexor muscles of forearm on left. Could grasp lightly with both hands; right stronger. Muscles of chest and abdomen not apparently affected, though respiration was not deep. Could raise neither leg from the bed,—in fact all motions of the thighs were lost. Flexion and extension of ankles and toes possible but weak. No paralysis of sphincter ani.

Reflexes: Triceps, quadriceps and Achilles absent. Abdominals and cremasterics lost. Chin and orbicular present. Muscular irritability to mechanical stimuli generally marked.

Sensation: No anesthesia to touch, pain or temperature anywhere. Slight general hyperesthesia, especially of legs, but tenderness was not marked over particular nerves or muscles.

Patient mentally clear. Speech normal. Urination had not been voluntary since the first catheterization. The diagnosis was given as acute anterior poliomyelitis. Prognosis, as to life, good; as to muscular efficiency, poor.

On September 11th the patient was seen again. Within two days after the onset of the paralysis the temperature had dropped to normal, the muscles of the neck had relaxed and the patient began to void urine voluntarily. Since that time there had been slight, gradual improvement in the use of certain muscles of the limbs. He had been on general supportive treatment.

Examination at this time showed that the patient had lost much flesh and that the extremities were somewhat atrophied. Skin, especially of hands, dry and scaly. Reflexes and sensation unchanged from former examination. Heart—systolic soft murmur at base and in neck. Sounds clear at apex. Lungs normal. Patient could now raise the right shoulder slightly and could flex and extend the right arm. Movements of left shoulder, and arm extension, impossible. Flexion of legs lost. Extension weak. Adduction and abduction weak. Outward rotation of right leg possible; of left lost. Movements of toes and ankles practically normal. No toe or wrist drop.

Electrical examination (considerably hampered by failure to use a strong current). To a moderate faradic current all hand and arm muscles contracted except the left triceps. The pectorals and deltoids failed to react to a painful current. The ulnar nerve conduction was normal. Legs—both quadriceps, sartorii and adductors failed to react to a painful current. All muscles below the knees reacted to a fairly strong current. To the galvanic current no muscles showed any increase of irritability and the cathode closure gave a stronger contraction than the anode.

The urine now contained much pus and some blood cells. The patient's appetite was good and his bowels regular. The cystitis was receiving active treatment. Massage was recommended, especially for the extremities.

For two months the patient was not seen again. He had been having massage and, while his strength had but slightly improved, when put in a wheel chair, he had managed to work himself around the room. The cystitis had persisted.

November 11th, 10 p.m. The patient was found reclining in a cushioned wheel chair. Face cyanotic. Rattle in bronchi audible across the room. Accessory respiratory muscles in action. Heart rate 136. Sounds booming; no murmur. Coarse mucous râles all over lungs in front. Bronchial respiration in right axilla and lower front. Heart sounds transmitted loudly through this area. Patient could move the right arm fairly readily and reach above his head. No paralysis of either forearm, though there was considerable weakness. On account of the critical condition of the patient the leg movements were not examined. Oxygen was sent for but before its arrival the dyspnea became extreme. The pulse went to 150 and was irregular. Oxygen administered for three minutes gave very marked relief and within an hour he was resting fairly well in bed, the pulse having dropped to 124. The regular stimulating and supportive treatment for pneumonia was instituted. Next day the whole right chest was flat to percussion, with bronchial respiration. There were coarse râles, numerous on the left. During the night dyspnea and weakness became extreme and the boy died at 5.30 a.m., November 13th, 1904.

The autopsy was performed by Dr. Burley and myself three hours post mortem. The spinal cord only was examined. Body considerably emaciated; muscular atrophy most marked about shoulders and arms, especially the left. Both legs markedly atrophic and about equal. Right chest, front and back, flat to percussion. The spinal cord was removed from the fourth cervical segment downward. Cerebro-spinal fluid clear and moderate in amount. Pial vessels very much injected. Cord somewhat smaller than normal. Consistency fairly firm except in the upper dorsal region, where it was rather soft. Transverse section of the fresh cord in the cervical, dorsal and lumbar region showed slight grayness of the white matter. This was most pronounced in the anterior columns and gradually faded toward the posterior parts of the lateral tracts. The gray matter in every section stood out prominently and was of a light pink color and showed considerable injection and softening in the anterior horn area.

Microscopical Examination.—All of the segments of the cord from the fourth cervical downward were examined. The stains employed were Nissl's methylene blue, hematoxylin and eosin, Weigert's medullated fiber stain, Weigert's neuroglia stain and Marchi's method for degenerated fibers. All of the segments

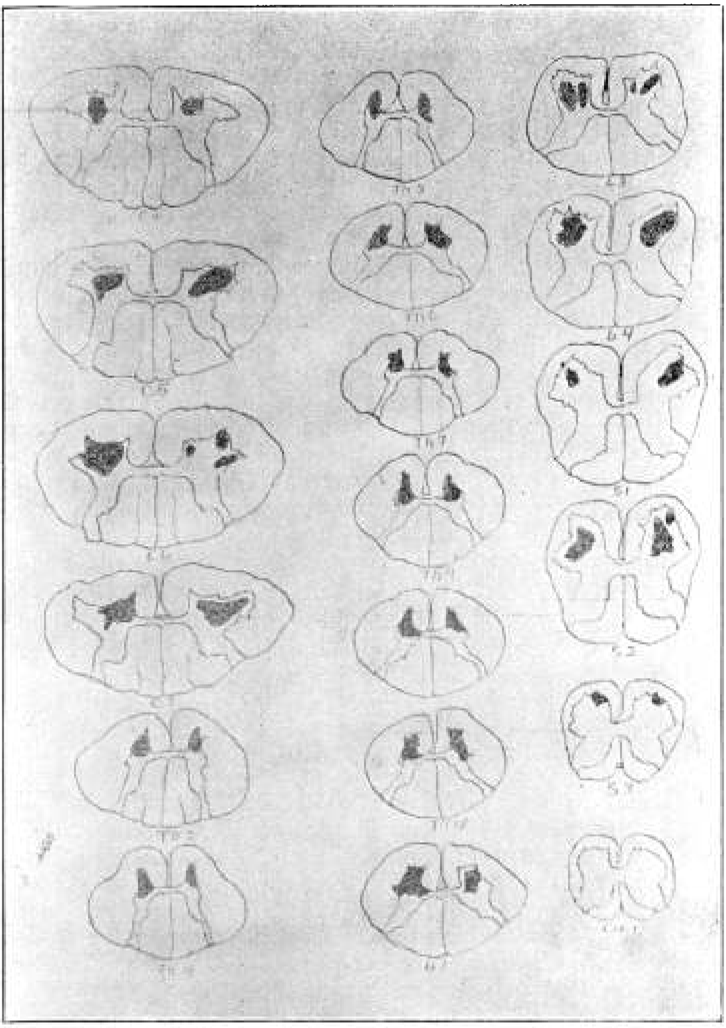

Fig. 1. Shaded portions represent the areas of destruction within the gray matter. Drawn by means of Edinger projection apparatus and photographed.

were extensively involved and, in order to prevent repetition, characteristic levels in the cervical, dorsal and lumbar segments only will be described, except where lesions show a marked departure from the common type.

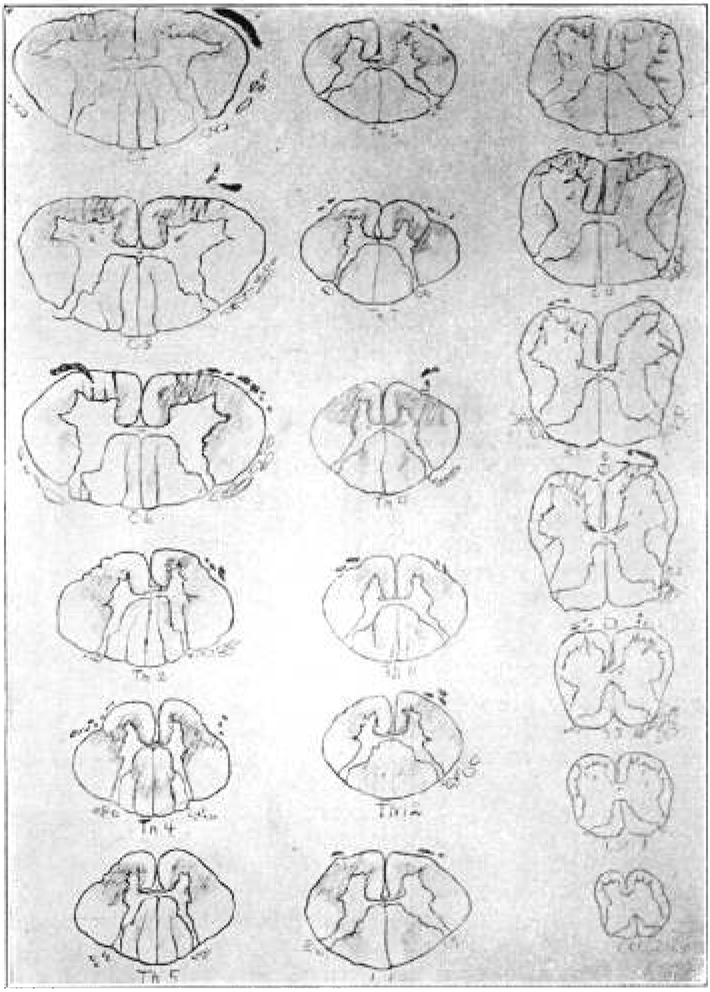

FIG 2. Shaded portions represent the areas of degeneration within the white matter. Drawn by means of Edinger projection apparatus and photographed.

The Cervical Cord.—The normal contour of the cord is lost.

The anterior horns are smaller than normal, causing a depression of the white matter over them. The disease process is bilateral, fairly symmetrical and limited in its active manifestations to the ventral gray matter. The anterior horns, however, are not wholly diseased but the lesions are distributed in irregular patches, which vary greatly at different levels. One group of cells may be involved in one segment and remain partially intact in another, while an entirely different area of the gray matter is affected.

Sections stained by Nissl's method show the following characteristics. With slight magnification the anterior horns can be identified only by the presence of very faintly staining cells within the gray matter, more especially near the periphery (hypertrophied neuroglia cells). A few remnants of nerve cells of the anterio-median group exist; in other levels a few cells remain in the antero-lateral group. The cells showing the least degree of involvement are in the intermedio-lateral group. In some sections no nerve cells can be found in the anterior horns and in general one is impressed with their great scarcity. Those that remain are coarsely granular and stain deeply. The chromatin appears heaped up in clumps and but little non-stainable substance is seen. The nuclei, when present, are round and located centrally and the nucleoli stain deeply. The cells are small, angular, flattened, long and spindle-shaped. Their prolongations appear suddenly broken off. No cells show any evidence of a chromatolysis. There is extensive hypertrophy and hyperplasia of neuroglia cells within the ventral gray matter. These cells are most numerous at the junction of the gray and white matter and are widely scattered throughout the body of the anterior horn. A few isolated hypertrophied neuroglia cells are seen in the periphery of the antero-lateral tracts in the white matter. A slight neuroglial proliferation is also evident near the base of the posterior horn. These hypertrophied neuroglia cells are large, stellate, and appear drawn out in places. The cytoplasm takes a faint bluish pink stain. The nucleus is large, pale and eccentric. Scattered among these cells are numerous normal glia nuclei. Specimens stained by the neuroglia method show a large increase in neuroglia fibers throughout the entire ventral gray matter, the anterior commissure and anterior root zone. The cytoplasm of the hypertrophied neuroglia cells does not stain by this method. The fibrils are short, coarse, straight and angular. They stain deeply, but seem to end abruptly, near the cell body, and at their periphery. Occasionally thin fibrils appear to mark out the boundary of the cells. Cells of this type are numerous in the immediate vicinity of the most pronounced vascular changes.

The blood vessels of the ventral gray matter show extensive changes. They are engorged with red blood cells and the walls are thickened. Many new vessels have been formed and in places the nervous elements are entirely replaced by a fibrillar network and distended capillaries. Branches of the anterior spinal artery and of the antero-lateral pial vessels are about equally affected. These vessels appear to lead directly into the patches of destruction within the gray matter, which may be considered partly as areas of anemic necrosis. These patches have a loose areolar structure and are made up of a fibrillar scar tissue with vascular proliferation. Nerve cells, nerve fibers and glia cells are absent, but newly-formed vessels are abundant. Occasionally these areas are crowded with polymorphonuclear leukocytes and small lymphocytes, but no foci are found which could be considered purulent.

The perivascular spaces of the pial and anterior spinal arteries leading toward these lesions are crowded with extravasated cells of various types, but only after they enter the gray matter.

Polymorphonuclear leukocytes are scattered throughout the areas of infiltration in small number. Small lymphocytes are closely packed about the larger vessels within the gray matter, four or five layers in depth. Within these areas a few isolated plasma cells are seen. The smaller capillaries within, and immediately surrounding the foci of destruction, show numerous characteristic plasma cells surrounding them. These cells have at times a mosaic-like arrangement and are usually oblong and angular with the characteristic appearance of the pale and eccentric nucleus and a heaping up of granules in the periphery. The larger and thicker vessels within the same areas are surrounded by an entirely different type of cell. A few plasm cells are present, but the prevailing type is a cell much larger than the plasma cell. It is round or oval in shape with a large, round, glassy nucleus, which shows a few deeply-staining granules The cytoplasm of the cell is very pale, and contains indistinct fine granules which are widely scattered. Occasionally slightly larger and irregular granules may be made out. By the Nissl method these granules appear of a purplish tint. Marchi's method shows the majority of them, at least, to be made up of fat or myelin detritus. Occasionally the cytoplasm has a vacuolated appearance and at times a cell can be seen with an inclusion of lymphocytes; in one instance two lymphocytes were included within a cell. These cells are likewise found scattered throughout the gray matter. They are probably of the compound granular type, called by some "granule" cells; by others "fat granule" cells. The vessels within the pia and anterior fissure are free from infiltration and evidence of thrombosis or embolism is nowhere present. The anterior spinal artery is never involved until it penetrates into the gray matter; but the pial vessels show a slight perivascular lymphocytic infiltration within the white matter, increasing in intensity as they near the gray matter. In some sections the anterior spinal branches appear normal and the patches of necrosis and scar tissue seem wholly dependent upon the disease of the peripheral vessels. Weigert's medullated fiber method shows the following condition:

There is distinct grayness of the anterior roots and a scarcity of fibers in the anterior root zones. The connective tissue is increased, especially near the gray matter. There are numerous large vacuoles within the antero-lateral white matter. The nerve fibers which remain are for the most part normal in size, but there are many smaller, and some very much larger ones. There is a marked scarcity of fibers in the anterior horns, especially in the antero-median part. The fibers which remain in the gray matter are thick, short and varicose. There is an extensive disappearance of fibers in the anterior commissure. The fibers entering the posterior horns have a normal appearance.

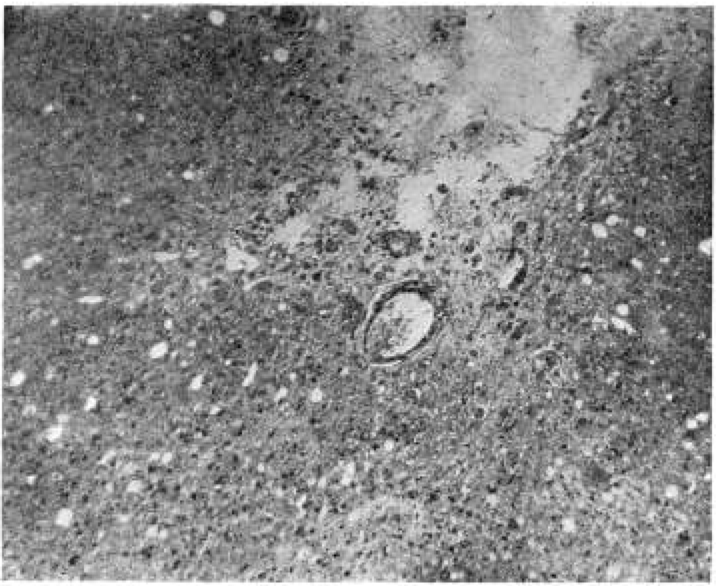

FIG. 3. Marchi specimen Dilated peripheral vessel surrounded by cells with black granules. Disintegration of gray matter.

Marchi specimens show numerous black degenerated fibers in the anterior commissure, some within the gray matter, many throughout the anterior root zone, and all the white matter adjacent to the gray matter. Scattered degenerated fibers are seen in the lateral parts of the posterior columns. Most of the larger cells in the perivascular spaces, and scattered throughout the gray matter, show large numbers of fine and coarse fat or myelin granules.

The Thoracic Cord.—The anterior horns throughout the entire thoracic cord are extensively atrophied and completely devoid of nerve cells or nerve fibers. The diameter of the cord is greatly diminished. The gray matter of both anterior horns is entirely replaced by a highly vascular and loosely reticular scar tissue, the interstices of which are in some areas closely filled with polymorphonuclear leukocytes. A few fairly normal cells may be seen at times in the intermedio-lateral group. The vascular changes are as pronounced as in the cervical region and involve about equally the branches of the anterior spinal artery and the peripheral vessels. In some sections the alterations in the branches of the pial vessels predominate; in others, changes about the anterior spinal artery are most extensive. On the whole, the former appear to share in the inflammatory process to the greatest extent.

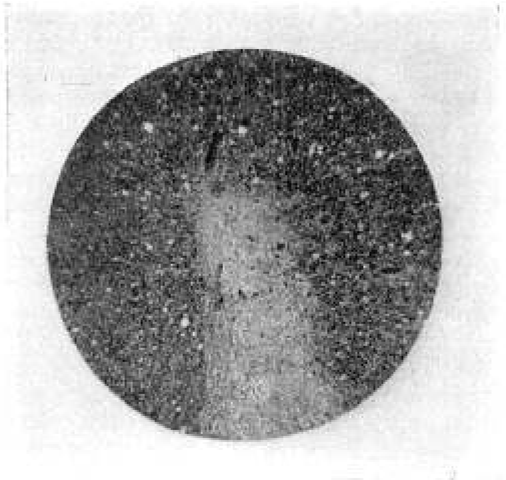

FIG. 4. Anterior horn of thoracic cord made up entirely of scar tissue and vessels.

A few hypertrophic neuroglia cells are seen, chiefly at the junction of the normal gray matter with the posterior horns. Nissl specimens demonstrate numerous cellular elements within the interstices of the scar tissue replacing the gray matter of the anterior horn. Many of these cells are plasma cells; some are of the compound granular type, while the majority are neuroglia cells and lymphocytes. A few isolated neuroglia fibers are seen within the destroyed areas. Where nerve fibers are present, they are short, coarse, stubby and varicose. The most absolute destruction of the ventral gray matter is at the level of the fifth thoracic segment.

The anterior roots are almost completely degenerated and the zone of white matter surrounding the anterior horns shows a great scarcity of nerve fibers. Only a few fibers can be seen in the anterior commissure. A similar, though rather less diffuse, area of degenerated nerve fibers is seen in the lateral parts of the posterior columns.

The cells of Clarke's column appear normal in number, but show distention, central chromatolysis, and a heaping up of granules in the periphery. The nuclei are eccentric. The network of fibers within Clarke's column is rather diminished in the upper part of the thoracic region.

The Lumbar Cord.—The destruction of the ventral gray matter is not as complete as in the thoracic region but has the same characteristic patchy arrangement that is seen in the cervical region. A few nerve cells in various stages of destruction are seen in the anterior horns; the remainder of the anterior horns is made up of numerous small deeply-staining neuroglia cells and many large, pale, hypertrophic neuroglia cells, especially in the vicinity of the larger vessels. These cells are also very numerous in the region of the anterior commissure. Few cells are seen in Clarke's column, and these have a normal appearance. The vessels entering the anterior horns from the periphery, especially with the anterior roots, show extensive alterations; some are very much thickened and surrounded by varying degrees of cellular infiltration. These cells are lymphocytes, closely crowded together. Aside from them there are a few scattered cells which answer the description of Unna's plasma cells, and some compound granular cells. Small clusters of nerve cells, which are small and irregular, are seen in the intermedio-lateral part of the anterior horn. The posterior horns are normal. Hematoxylin and eosin specimens show the anterior horns to be made up of a fine fibrillar network, and numerous large neuroglia cells with pale pink protoplasm.

Marchi specimens show degenerated areas corresponding to those in the cervical and thoracic region, but much milder in degree. There is an enormous new vascular formation within the anterior horns, especially in the antero-lateral part. The branches of the anterior spinal artery are somewhat distended, but the greater part of the vascular changes are derived from branches of the pial vessels. Minute hemorrhages and extensive perivascular infiltration are seen. The pia is slightly thickened. There is an extensive neuroglial proliferation in the anterior commissure and in the middle part of the gray matter. A few scattered neuroglia fibers are seen in the anterior horn where the destruction is most pronounced. Neuroglia cells are increased at the junction of the gray and white matter, in the white matter of the anterior root zone and in the periphery of the cord from the posterior horns forward.

Weigert's stain shows a great diminution of fibers in the gray matter surrounding the anterior horn. The anterior commissure shows more normal fibers than previous sections.

In the fourth lumbar segment many normal cells are seen in the antero-mesial part of the gray matter, and the foci of destruction are smaller and nearer the periphery. The vascular changes are confined almost wholly to the peripheral vessels.

Sections from the sacral segments show similar changes, though to a less degree. Again the vessels entering from the periphery, especially with the anterior roots, show the most extensive inflammatory changes. The foci of destruction are small and many normal nerve cells are present. The same hyperplasia and hypertrophy of neuroglia cells is present near the foci. The white matter shows no involvement.

A bacteriological examination was not undertaken, but no micro-organisms could be demonstrated in any of the sections.

Summary.—Clinically we are dealing with a strong and apparently healthy boy who, after a few days of general indisposition, suddenly developed a flaccid paralysis beginning in the legs and within 24 hours spreading to the arms. These phenomena were accompanied by a rise in temperature. After a brief, acute stage, gradual but slight improvement took place. After 13 weeks, death ensued from pneumonia. The onset was sudden. There was no history of preceding infectious disease or of exposure. As far as I am able to learn, no other similar cases occurred in Worcester at this time, neither were there any epidemics of cerebro-spinal meningitis or other infectious diseases. The presence of tubercular disease in a brother and sister is highly suggestive in view of the absence of any definite etiological factors, but microscopical examination failed to show any micro-organisms.

In summing up the anatomical changes we find throughout extensive alteration of the gray matter of the cord, more especially of the anterior horns. The posterior horns have suffered less although here and there slight vascular changes, as well as a mild neuroglial proliferation, are evident. The entire cord shows primarily a widespread inflammatory condition of the gray matter. The vessels are thickened, distended, engorged with red blood cells and increased in number. The perivascular spaces are crowded with cells, chiefly of the lymphoid variety. Plasma cells are fairly common and most abundant about the thickest vessels and around minute newly-formed vessels. In these regions there are but few lymphoid cells and the tissue destruction is most pronounced. The areas in which the lymphoid cells seem to predominate appear to be of more recent origin. They are also numerous in areas immediately surrounding the vessels as they enter the gray matter, becoming less intense as the destroyed areas are reached. Compound granular cells are found in some sections in fairly large numbers about the blood vessels and within the surrounding tissue. Large areas of extravasated leukocytes are found in the upper dorsal region, almost replacing the entire gray matter in some levels. Similar smaller foci are present in the cervical and lumbar regions, but no pus or abscess cavities are found. None of the vessels show any evidence of thrombosis or embolism. The vessels which participate in the changes are derived both from the anterior spinal artery and from the pia. The largest foci are found in the course of the branches of the anterior spinal in the cervical region, but the pial vessels show more involvement in the lower thoracic and lumbar cord. The perivascular infiltration is confined almost entirely to the gray matter though some of the vessels which enter from the periphery show a moderate degree of infiltration within the white matter, but increasing in intensity as they near the gray matter. In every section the anterior spinal artery appears normal until it penetrates into the gray matter.

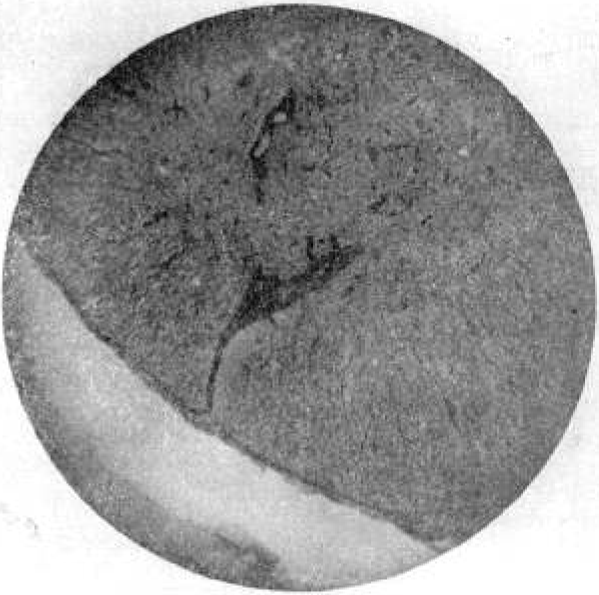

FIG. 5. Pial vessels showing extensive perivascular infiltration. Many newly-formed, thickened vessels in the gray matter.

The anterior horns are almost entirely devoid of nerve cells and fibers. Those cells which remain are seen chiefly in the extreme periphery of the gray matter, but in different areas at the different levels, apparently depending directly upon the blood supply. They have undergone great destructive changes; are smaller than normal; the processes appear suddenly broken off; the Nissl granules are arranged in large, coarse, deeply-staining clumps; the cells are angular, flat, narrow or spindle-shaped, as if they had been subjected to mechanical pressure. Very seldom a normal cell is seen while the majority are mere remnants. The group of cells which are most constant are the intermedio-lateral. In the thoracic region the anterior horns are made up almost entirely of scar tissue, very rich in vessels and without any nerve cells. Cells of Clarke's columns are not diminished in number but show the axonal reaction. The anterior horns throughout are atrophied, more especially in the dorsal region, causing a distinct flattening of the cord over them. The destruction is bilateral with practically complete involvement of the anterior horns of the dorsal region but with a peculiar destruction in small and large irregular patches in the cervical and lumbar regions.

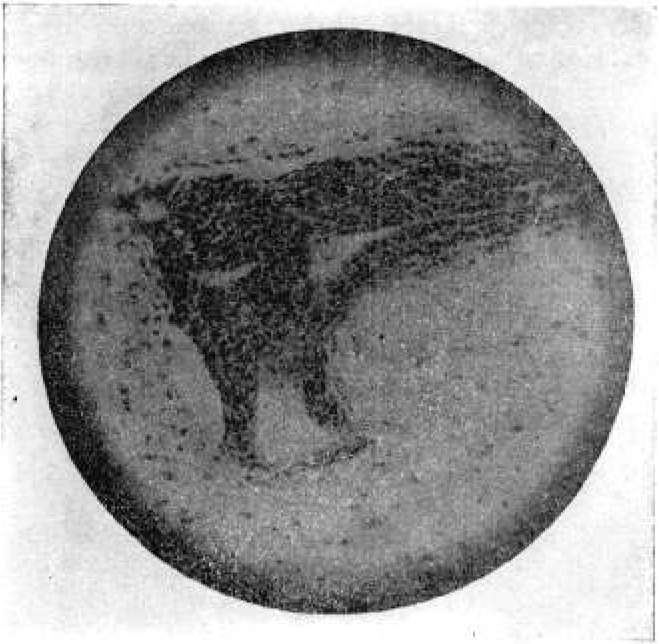

FIG. 6. Same vessel as that pictured, under higher power.

(*To be continued*)